

RIGHT SIDED AORTA ASSOCIATED WITH TRACHEO-OESOPHAGEAL FISTULA AND OESOPHAGEAL ATRESIA

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RIGHT sided aorta is a known association with tracheo-oesophageal fistula. In the Royal Belfast Hospital for Sick Children we would expect to see approximately three cases in ten years. The cases reported all presented during a 12 week period in 1981.

Case 1

Born at 40 weeks gestation, weight 2.46 kg. Abnormalities-TOF (proximal atresia with distal fistula). TOF was repaired by ligation of the fistula and single layer interrupted nylon anastomosis of the proximal pouch and distal oesophagus on day one via a right thoracotomy at which a right sided aorta was recognised. Cardiac catheterization in the post-operative period showed right sided aortic arch with residual left aortic arch giving rise to the left subclavian artery, coarctation of right aortic arch with narrowing of anomalous right subclavian artery arising at the level of T4 from the right aortic arch.

The child's IVP showed normal renal tracts and bladder. Vomiting in the first few weeks post-operatively was due to partial thoracic stomach demonstrated by barium swallow.

The child is presently alive and well; and has had no cardiac surgery to date.

Case 2

Born at 34 weeks' gestation, weight 2.04 kg. Abnormalities-TOF (proximal atresia with distal fistula). The TOF was repaired as in case 1 via right thoracotomy on day one. At operation a right sided aortic arch with anomalous right subclavian artery arising at the level of T4 was recognised. Location of the tracheal fistula and distal oesophagus was difficult and required full mobilisation of the thoracic aorta which was retracted on tapes. Despite care not to do so aortic retraction resulted in poor distal flow and subsequent acute renal tubular necrosis. In addition, the femoral pulses were (persistently) weak post-operatively having been pronounced pre-operatively. Following this episode the child did not recover. At post mortem a right sided aorta with anomalous right subclavian artery and patent ductus arteriosus were confirmed. The descending aorta showed intimal damage.

Case 3

Born at 40 weeks' weight 2.26 kg. Abnormalities-TOF (proximal atresia with distal fistula) The TOF was repaired as in case one via right thoracotomy on day one at which a right sided aortic arch was noted. Anastomosis was not possible as the upper pouch was very high and the lower oesophagus was displaced by the right aorta. The oesophageal components were approximated with a view to reconstruction at a later date.

Cardiac catheterization in the post-operative period showed right sided aortic arch with ventricular septal defect and overriding aorta. Pulmonary valve atresia, atresia of the pulmonary artery with hypoplasia of the right and left branches of the pulmonary artery was also noted along with patent ductus arteriosus and persistent left superior vena cava.

The child acquired fatal septicaemia at four weeks.

COMMENTS

The incidence of cardiac anomalies associated with TOF is approximately 20-25 per cent half of which are minor abnormalities which do not interfere with the child's prognosis,¹ such a right sided aortic arch, anomalous right subclavian artery and persistent left superior vena cava. A further breakdown demonstrates that

cardiac anomalies are three times more common in cases with additional non-cardiac malformations² when compared with isolated TOF and twenty-five times more common than the incidence of anomalous cardiac development in the general population.² The incidence of right sided aorta in association with oesophageal atresia is approximately 4 per cent^{1,3} as opposed to 0.1 per cent in the population at large.⁴ There is a similar incidence of anomalous right subclavian artery and persistent left superior vena cava with TOF,^{1,3} and these lesions along with right sided aorta are mostly of interest to the surgeon during dissection of the chest, as mobilization of the TOF and the lower oesophagus in particular, is much more difficult, as the right sided aorta occupies the position normally taken up by that structure and displaces it towards the midline and left chest. Anastomosis of the upper and lower pouch is thus more difficult and if an anomalous right subclavian artery is also present not only does the upper pouch tend to be shorter than normal, but the course of the newly formed oesophagus will be longer to circumvent these vessels.

For the child the association of right sided aorta with Fallot's tetralogy (14-34 per cent) and truncus arteriosus (12-36 per cent)⁴ is more significant than the oesophageal pathology as in all major series the life expectancy is reduced by 50 per cent when major cardiac anomalies are present.¹⁻⁴

Also co-existing third and fourth pouch anomalies may be present. Of particular interest are thymic and parathyroid anomalies which may be as profound as complete aplasia.⁵ None of the cases reported here demonstrated any abnormality in calcium metabolism and thymic function was investigated only in one case, with no apparent abnormality.

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